World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

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Case report of *Paraproteinemia*

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World Journal of Advanced Research and Reviews, 2022, 16(02), 848–850

Publication history: Received on 26 July 20212 revised on 18 November 2022; accepted on 20 November 2022

Article DOI: https://doi.org/10.30574/wjarr.2022.16.2.0771

Abstract

Different peripheral neuropathies are linked to paraproteinemia. Anti-myelin-associated glycoprotein (MAG) neuropathy, immunoglobulin or its fragment deposition in the interstitium, immunoglobulin light chain amyloidosis (AL amyloidosis) and paraneoplastic mechanisms that cannot be solely attributed to immunoglobulin or its fragment deposition, represented by polyneuropathy, organotypic, are the three main causes of neuropathy correlated with paraproteinemia. Nerve conduction metrics slow down in patients with POEMS syndrome and anti-MAG neuropathy. An approximately (3.2%) of people over the age of 45-50 have paraproteins in their bodies. Plasma cell diseases can be thought of as a continuum of illnesses ranging from asymptomatic myeloma to symptomatic monoclonal gammopathy of unknown significance (MGUS). A low amount of paraprotein (30 g/L), a low percentage of plasma cells in the bone marrow, and the absence of organ or tissue damage from myeloma are the characteristics of MGUS (predominantly renal, skeletal or bone marrow impairment.) No treatment is necessary for MGUS, and there is a 1% annual risk.

Keywords: Paraproteinemia; POEMS; anti-MAG neuropathy; MGUS

1. Introduction

A diverse category of diseases known as paraproteinemias are characterized by the proliferation of monoclonal proteins by monoclonal plasma cells [2]. Clinicians should be aware of them because neuropathies frequently co-occur with them. When neuropathy is the presenting feature, neurologists have a special responsibility because they may find clinical, laboratory, radiologic, electrodiagnostic, or biopsy findings that may identify the underlying paraproteinemia [1-3]. Between the various paraproteinemias, there are considerable differences in the prevalence of neuropathies in these patients and the degree to which they predominate the clinical picture. Treatments may target the neuropathy specifically or the underlying hematologic condition [4]. The neurologist and haematologist can collaborate on developing treatment plans and objectives for all paraproteinemias patients, and they can also give follow-up and monitoring to see how the neuropathy is responding to the medication [2].

2. Case -1

A 60 years old gentleman, retired officer by the Government Public sector services, holding a bachelor's degree in Engineering. He visited the Neurology clinic with his son during the last quarter of the year 2021 with chief complaints of weakness and wasting of proximal greater than distal upper limbs started on right followed by left after 1 month for the past 6 months. The patient was right-handed and had no history of lower limb weakness, fasciculation, stiffness, neck pain, or radiating pain. Moreover, no history of disturbances in upper or lower limb cranial nerve symptoms, gait abnormality, bowel bladder involvement trauma, weight loss or loss of appetite was reported. The patient was normotensive with no known diabetes condition with no significant family and personal h/o .previously he was treated for intravenous MP for five days, which failed to show meaningful improvement. At this point, differential diagnosis of

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bi brachial amyotrophy motor neuron disease. MMN –CB and bilateral pan brachial peixitis were considered. The patient was moderately built and nourished general examination revealed no abnormalities findings pulse 80/min, BP 126/82 mm/hg, and RR 12/min. His mini-mental state examination (MMSE) and cranial nerve function examination were reported to be normal. Motor function examination demonstrated bilateral severe wasting of arm and forearm muscles, hypotonia in both upper limbs, and 1/5 muscle power in supraspinatus, infraspinatus, deltoid, latissimus dorsi, pectoralis serratus anterior, rhomboid, biceps, triceps, forearm muscles, small muscles of hand with very weak hand grip. Muscle tone and power (5/5) were normal in lower limb muscles. Deep tendon reflexes were absent in biceps, triceps and supinator, knee and ankle reflexes were reported to be one plus, Bilateral planters -flexor. On MRI imaging, mild diffuse disc bulges with disc osteophyte complexes reduced disc space and end plate irregularities at C_4/C_5, C_5/C_6, and C_6/C_7 levels, with no associated spinal canal or neural foraminal narrowing, were observed. There was diffuse disc desiccation with mild diffuse disc bulges at L1/1,2, L_2/1,3 and L_3/L4 levels, with no associated spinal canal or neural foraminal narrowing. Anterior and posterior osteophytes were observed with reduced disc spaces and end plate irregularities and posterior marginal osteophytes, reduced disc spaces and end plate irregularities at these levels along with mild dorsolumbar kyphosis. Bilateral facet joint degenerative arthropathy was noted at L_1/L_2, L_2/L_3, L3/L_4,L_4/L_5 and L_5/S_1 levels. Vertebral body heights and alignment were normal. Marrow signal intensity was also reported to be normal. The rest of the intervertebral discs showed a normal intensity pattern with a normal craniovertebral junction. MRI brachial plexus demonstrated bilateral plexitis. Electromyography showed the presence of fibrillations; positive sharp waves, complex repetitive discharges, and fasciculation in both upper limbs with a long duration large. Amplitude polyphasic motor until action potential (MUAPS), in paraspinal thorasic muscles, CRD are seen with normal lower limb, the whole body position emission tomography produced abnormal findings and the urine Bence Jones protein test was negative. Basing these findings, the diagnosis of bi- brachial amyotrophy with motor neuron disease and bilateral plan brachial plexitis was made, considering the monoclonal gammopathy or neurological significance as the causative etiology.

3. Case -2

A 56 years old male patient presented with tingling, and paraesthesia followed by hands for the past 4 months. He also experienced a history of weakness in lower and upper limbs 3 and 2 months before respectively along with swelling on the right side of the neck for the past 5-6 years. Non-tender, firm, cervical Lymphadenopathy of 2x2 cm was observed on the left side. Mini-mental state examination score was 30/30. Cranial nerve function was found to be normal, mildly reduced distal muscle tone was reported in upper and lower limbs. Muscle power was 5/5 in the shoulder, 5/5 elbow, 3/5 in the wrist,4/5 in the knee, 3/5 in the ankle, and small muscles of the hand were weak. All deep tendon reflexes were absent. Sensory examination showed normal findings of touch, pinrick, and joint positions in all 4 limbs. In bilateral lower limbs, impaired vibration sense was reported up to tibial tuberosity, and cerebella functions were normal. Biochemical investigations(CBC, LFT, RFT, paraneoplastic profile, urine Bence Jones, serum immunofixation, bonemarrow biopsy, neuropathy involving all four limbs, CT of the chest was normal; CT abdomen showed liver hemangioma and splenomegaly. Neck Ultrasonography showed enlargement of levels ii, iii, and iv group lymph nodes on the right side and sub-centrimetric lymph nodes on the right side. These findings led to the diagnosis of multicentric Castleman's disease presenting as peripheral neuropathy.

4. Conclusion

Therapeutic plasmapheresis problems specific to paraproteins are uncommon. Therapeutic plasmapheresis has increased the therapeutic options available for managing paraproteinemia as an adjunctive treatment.

Compliance with ethical standards

Acknowledgments

The author acknowledged to the key officers of PMSSY, BMCR & I, Bengalore.

Disclosure of conflict of interest

There are no conflicts of interests with any other funding agencies.

Statement of informed consent

Institutional permission and consent obtained from the patients and caregivers

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